

THE
TREATMENT OF INFANTILE
SPASTIC PARALYSIS.

BY

ROBERT JONES, F.R.C.S.E.

SURGEON, ROYAL SOUTHERN HOSPITAL; AND LIVERPOOL HOSPITAL FOR
CHRONIC DISEASES OF CHILDREN, HESWALL, CHESHIRE.

(Reprinted from *Liverpool Medico-Chirurgical Journal*, October 1902.)

WELLCOME INSTITUTE LIBRARY	
Coll.	welMomec
Coll.	pam
No.	WC 555
	1 9 0 2
	J 7 9 t



22500654770

The Treatment of Infantile Spastic Paralysis.

By

ROBERT JONES, F.R.C.S.E.,

Surgeon, Royal Southern Hospital, and Liverpool Hospital
for Chronic Diseases of Children, Heswall, Cheshire.

I VENTURE to bring before you a subject which for many years has interested me and which until quite recently has so far as treatment is concerned, suffered unmerited neglect. There is something so uninviting and hopeless-looking about a case of cerebral diplegia that a man must possess in addition to a theory of attack, no small fund of enthusiasm before he decides on treatment. Cases of infantile hemiplegia, with half their brain and half their extremities in working condition, offer less obstacles to management if less scope for ingenuity.

It is impossible to enter fully into the subject of cerebral paralysis, but I can indicate certain clinical types which offer adequate reward to surgical endeavour, and point out the remedial methods which in my experience have proved of value.

For the purposes of this paper I would group cases into those of (1) Infantile Hemiplegia; (2) Cerebral Diplegia; and (3) Spastic Paraplegia.

Of 850 cases I have been able to collect from various sources, 510 were hemiplegic; 30 monoplegic; 142 paraplegic; 157 diplegic.

This vast disproportion between the hemiplegic and the other groups is not borne out in my own experience, which favours the preponderance of the diplegic type.

Most generally the hemiplegia is an acquired, not a congenital affection. It usually appears before the fourth year. The onset is very often heralded by convulsions, and in quite a number of cases there are acute febrile symptoms, which are in interesting contrast to the onset of adult hemiplegia.

From the first, the reflexes are exaggerated and the arm and limb are powerless. Deformities soon follow. The foot is arched and adducted; the knee is bent; the hip is internally rotated, somewhat adducted, and the femur is bent on the trunk. The arm is huddled to the body, the elbow is flexed to a right angle, the wrist is dropped, the hand pronated and deviates towards the ulnar side, and the fingers generally grasp the thumb, which lies very forcibly adducted. As in the case of adults, rigidity of the affected muscles comes on in the generality of cases. This rigidity becomes more marked if the limb be moved either passively or by the child, and disappears under an anæsthetic. The aphasia so often present is motor in character, and may accompany either left or right hemiplegia. Then follows an inequality of growth. This is more marked in the arm than in the leg; a fact probably accounted for by the difference in nutrition, due to the more frequent exercise of the leg. I have seen the affected arm three inches or more shortened; it is rare to find the leg over an inch shorter than its fellow.

Of chief interest to us from the point of view of treatment are the following facts:—

- (a) The upper limb is more severely affected than the lower.
- (b) The lesion of the upper limb is more permanent.
- (c) The power of dorsiflexion of the hand, with simultaneous extension of the fingers, is lost.

(d) That movements are performed without precision, spasmodically and slowly.

- (e) That the power of adduction of the thumb is often lost.

The disabilities of the lower limb are generally:—

- (a) Contraction of the knee.
- (b) Extension of the foot.
- (c) Internal rotation of the femur with adduction.
- (d) General rigidity.

The cerebral diplegic group is by far the most serious, as we have here to deal with both arms and feet. Unless the hands can be used, the surgeon is sorely handicapped in any

effort he may make to improve the condition of the limb. Clinically, we may divide this group into—

(a) Cases with and cases without severe mental complication.

(b) Complete and partial disability of hands.

(c) Complete or partial disability of limbs.

(d) Cases complicated by athetotic movements.

In a number of cases the spasm is confined to the limbs, and this is a group to which we give the name of Little's Disease, or Spastic Paraplegia. Of the vast majority of cases even in this last group a varying degree of mental derangement will be noted, and in many, athetotic movements are present. A typical case of spastic paraplegia brought to the surgeon at the age, say, of 12 months is characteristic; usually no contractions have occurred at the hip or knee; the child's legs are rigid; the toes are pointed; there is usually no internal rotation, and adduction is not sufficiently severe to cause a crossing of the limbs. The reflexes are exaggerated, the patellar reflex not usually causing a knee-jerk but a leg-jerk. If the little patient be held by its arms there is no endeavour made to separate the limbs; and should the toes be brought to the ground, and an effort made by the child to walk, both limbs move synchronously and in parallel lines. When passive separation of the limb is made, although it is easily effected, one can see and feel the antagonistic efforts of the powerful adductors. If at a later stage the patient is able to walk, several changes will be noted. The adduction will be more marked; the scissors walk will have developed, and a characteristic dragging of one knee round the other will be noted, which becomes more pronounced when any attempt at running be made. The body pressure is mainly transmitted to the ball of the toe. These contractures, however, at this stage, are generally spasmodic, and there is no appreciable shortening in muscle length. This shortening occurs at a still later stage, and is known as contracture.

In quite a number of cases the patients are in a hopeless

position as far as walking is concerned. Any effort they make to move only serves to throw the muscles into violent contractions and the legs into extreme adduction. The most severe type of contractions in the legs may be associated with but very slight mental defects, and unaccompanied by diplegia.

Before discussing treatment I will briefly touch upon the pathology of spasmodic paralysis, if only to suggest how futile are operative procedures directed to the primary lesion. The pathological conditions in hemiplegia, paraplegia, and diplegia are the same in kind. The symptoms are due to the retardation of growth, resulting generally from embolism or thrombosis, together with changes in the spinal cord. In later cases one finds wasting and sclerosis of the motor tracts, with often a loss of substance in the form of cavities or cysts, known as porencephalus. These cysts occur on the surface of the brain, and sometimes dip fairly deeply into it. They seem to be a late result in a growing brain, and to have produced an extensive scar substitute for cerebral tissue. Should the porencephalus or sclerosis be unilateral, hemiplegia results. If the scar is bilateral, diplegia or spastic paralysis ensues. The lesions, therefore, are a late product of a hæmorrhage, an embolism or a localised encephalitis. In the cord, degenerations of the pyramidal tract or the lateral columns are to be found. Sachs and Peterson have analysed 73 autopsies and found the following conditions :—

Atrophy, sclerosis and cysts,	40
Hæmorrhage,	20
Embolism,	7
Thrombosis,	5
Tubercle,	1
	—
	73

Osler, in an examination of 90 brains, found a vascular lesion in 16,—7 due to hæmorrhage, 9 to embolism.

The treatment of spastic paralysis has been too long in the hands of the physician for much real progress to have been

made. Indeed from medicine in this affection we have nothing to expect apart from very indirect results, and we have only to scan the text-books on neurology to realise the note of pessimism which is sounded. Even Sir William Gowers, in his *Diseases of the Nervous System*, says, "The tendo Achillis is sometimes divided for contraction of the calf muscles in infantile spastic paralysis, but the operation is useless and ought never to be performed." The same opinion has been pronounced by other distinguished men, so one can see how surgery has been silenced in the matter. I would argue that a large proportion of children suffering from severe spastic paralysis may be transformed into useful members of the community, improved both in body and mind by surgical methods, enabled to walk with comparatively little deformity, many requiring only the aid to be derived from one or two sticks.

The class of case which we can place outside remedial art is the idiot, the microcephalic, and the violent, irritable type of diplegic, so often seen, subject to fits and active athetotic movements, who has generally lost all control over his secretions. The treatment of any condition short of this may be undertaken with varying success, subject to conditions which obtain in any surgical case requiring prolonged attention. For instance, active treatment may be needed for nearly two years. It would therefore be unwise to admit a case into hospital for two months and then send it to a miserable home, where neglect would be the inevitable sequence. Such a case, however, after hospital treatment, secure in the care of anxious intelligent parents, no matter how poor, would prove a credit to all concerned. These are important matters which the surgeon must consider before he undertakes his work. Another class which gives the greatest anxiety and trouble is that where the affection of the hands is of such a kind as to promise but slight hope of their assistance to the limbs during walking. Before despairing, however, I think it is well to give such hands the opportunity of a careful trial, both as a mental discipline and because success sometimes exceeds expectation.

I would divide the treatment of all cases of spastic paralysis into operative and post-operative; for although mechanism is involved in nearly every case, there is hardly a case which we are called upon to treat without invoking operative aid.

Infantile hemiplegia usually affects the arm much more than it does the leg. This is almost invariably the case, and in this particular it differs from diplegia, where, when the four limbs are attacked, the hands are frequently less severely affected than the limbs. Indeed, in hemiplegia the paralysis of the hand is sometimes absolute, and in addition we have a complication in the shape of rigidity. The behaviour of the lower limb differs also from that of spastic paraplegia, in that the adductor spasm is proportionately not so marked.

The treatment of the hand and arm in infantile hemiplegia is distinctly less promising than in the diplegic case, but there are clinical signs to which I would draw your attention, which help us to prognose success or failure. If the paralysis is complete, or, in other words, if the little patient is never known to relax his spasm, treatment is futile. If he only moves the fingers of his affected hand in conjunction with the fingers of the opposite side, the results will in all probability be discouraging. In all cases where the parents are able to truly say, in the spirit of true observers, that the patient is able to do more with the hand now than he was a little while ago, the success of treatment is assured. Similarly, where any degree of voluntary relaxation of spasm exists apart from an associated movement on the opposite side, treatment is emphatically indicated.

Noting that the dominant deformity in both hand and elbow is pronation and carpal flexion, treatment should consist in fixing the elbow supine and in fully extending the wrist. The full extension of the wrist should be combined with that of the fingers, and a special arrangement adapted to keep the thumb at right angles to the palm. The spasm in these cases is often so pronounced that the extension of the wrist and fingers must be brought about very gradually. If the elbow

is affected by contracture of the biceps and brachialis anticus, supination may be combined with extension. If this be not the case, the flexed position of the elbow will suffice. If, instead of being firmly pronated, the elbow lies semi-proned, it is not necessary to treat it, and all one's energies should be directed to the hand. It is difficult to give a reason as to how improvement comes about, but it may be taken as an axiom that prolonged fixation of spastic muscles in a position opposed to their contraction lessens the severity of the spasm. This is true wherever spasm may be found, and its influence may be tested even in spasmodic torticollis, intractable as we know that affection to be. It would appear as if the group of muscles at last got tired of trying to pull. This treatment of full extension may be discarded in about twelve months if during the whole of that period it has been kept up without intermission. The test for relaxation must be the power of voluntary movement, however slow it may be. It will be noted that generally at this stage the patient, in endeavouring to extend his wrist, will first of all close his fingers, and will only open them on completion of extension. The process is reversed when the wrist is flexed. In order to meet this difficulty, the splint employed to extend both wrist and fingers is modified so as to extend the wrist alone and allow freedom to the fingers. At this stage, or earlier, the surgeon may decide whether in a given case success may be predicted, and if he is in doubt, operation should unhesitatingly be performed. The operative treatment will consist of tenotomy or tendon transplantation: myotomy need only be mentioned to be avoided. An incision is made over the tendon of the flexor carpi ulnaris just above the annular ligament, another is made over the flexor carpi radialis, and both tendons are divided low down and taken, (*a*) the flexor ulnaris to be inserted into the extensor ulnaris, and (*b*) the radial flexor into the radial extensor. I performed this operation for the first time some years ago upon two spastic children whom I showed before the Society for the Study of Diseases in Children, and in both

instances voluntary movements were steadily performed, and one, a girl of 9, was able to write quite a respectable hand. At the present time one of the worst cases of athetosis in connection with cerebral diplegia is an inmate of the Liverpool Country Hospital for Chronic Diseases of Children. When he entered the hospital nearly two years ago both limbs were firmly adducted, contracture had occurred, which had so affected the feet that plantar flexion had taken place with almost evulsion of each astragalus and cuboid, the skin of each dorsum being reddened by pressure from within. The hands were firmly flexed and but little voluntary movement existed. Athetotic movements, more resembling chorea, with occasional jerks which almost lifted the patient from his bed, complicated treatment. So far as the hands were concerned, hyper-extension such as I have described was practised, and later division of the flexors of the wrist. The little patient has steadily progressed. He can voluntarily move his limbs in all directions ; with aid he can take a few strides.

In order to overcome severe spasmodic pronation of the forearm, my friend Mr Tubby has changed the point of insertion from the front to the back of the radius of the pronator radii teres, thus transforming the muscle into a supinator. The operation has been several times successfully performed.

Tenotomy alone, has proved distinctly disappointing, although one has had an occasional successful case. The operation should be confined to the division of the flexor carpi radialis and ulnaris. It is, in my opinion, better to elongate the other flexors of the hand by a long median incision, such as one would employ in lengthening the tendo Achillis. Tendon transplantation, however, is a better operation, less complicated and more reliable. The surgeon's art, however, does not end with the operation, and over-extension of the wrist leaving the fingers free should be practised for a further few weeks. In order to prevent adhesions after the operation, the wrist should be freely but withal very gently moved in about a fortnight's time. Whether an operation has been performed or no,

the final stages of treatment are identical. They should consist in getting both guardian and patient to do all in their power to practise movements from simple to complex. The surgeon must inspire confidence and instil enthusiasm. This cannot be done by a long face which crushes hope, nor by doleful references to a still more doleful pathology. In most cities in England, neurology seems to make the physician a fatalist and one who clothes himself with gloom and pessimism. Of all departments in medicine this is surely the one where the brighter side of things should be kept in the foreground. Failure depends upon men as well as measures, and a thoughtless impatient word of discouragement may paralyse all effort.

The nature of the movements to be practised must be left to the ingenuity of the surgeon. The principles which should govern him may, however, be indicated here:—

(a) The movements should be practised slowly and without excitement.

(b) They should be made interesting to the child.

(c) Those opposed to the direction of deformity should predominate.

(d) Those presenting the greatest difficulty should be chiefly practised.

Just a word before we deal with paraplegia regarding tenotomy of the spastic muscle. Empiricism has taught us that, for some reason or another, tenotomy lessens, both in frequency and intensity, the spasmodic element in paraplegia. I do not merely mean to say that division of the tendo Achillis controls spasm in the calf muscles, although of course it does, but rather that spasm in which those muscles are not directly concerned is also influenced. This is beyond all question, and must have been noted by everybody who has had the opportunity of observing, and the fact has now reached the robust stage when physiological explanations are vouchsafed.

Whitman urges that by elongation of the tendon the response to the exaggerated motor impulses is lessened, and an opportunity for more effective control is afforded.

Mr Tubby has propounded the theory that once the immediate pathological effect of the central nervous lesion has subsided, the spinal cord remains in a state of undue reflex excitability. A tightly contracted muscle and tendon tend to augment this condition, and so induce in themselves further contractions. In other words, there is a vicious circle of reflex action which can be interrupted by section of the tendon, and diminution in the tension of the affected muscle and tendon.

Lorenz attributes the good effect to the shortening of the bellies of the tenotomised muscles, so that their range of action is diminished. Both these theories refer of course to spasm with which the divided muscle is concerned, and do not explain the diminution in spasm experienced elsewhere. We must further remember that the opponents of contracted muscles are always elongated and weak, and that the rest afforded them by tenotomies, by relieving them of strain, helps to restore muscular equilibrium.

The practical deduction from these observations is, that no opportunity should be lost of performing a tenotomy. Even in mild cases, where a spastic tendon is to be felt, let us ruthlessly divide it.

If the surgeon has decided that a case of spastic paraplegia is suitable for treatment, a splint should be prepared, so designed as to keep the limbs in marked abduction. The area over the hamstrings, the adductors at the groin, and the tendo Achillis should be prepared for operation. The adductors should be first attacked. An incision an inch and a half long should be made to the inside of the adductor longus. This muscle should be seized by a Spencer Wells or a small open forceps, and about three-quarters of an inch of it removed. The limb is then abducted, and portions of the adductor brevis and gracilis are excised in similar fashion. The horizontal portion of the adductor magnus, and if necessary the pectineus, is divided, and also any tissue, muscular or fibrous, obstructive to an absolutely free abduction of the

femur. Experience has shown me that although the chief offenders are the adductors longus and brevis, nevertheless the deeper muscles often require division. To anyone who has practised this operation, the futility of attempts to effectively divide the muscles subcutaneously will be apparent. Division is followed with but little hæmorrhage, and the wounds are closed without drainage. Having exsected the pieces of the adductors, each tendo Achillis is divided or lengthened, and rectangular splints are applied to the foot. The limbs are then well abducted, and the surgeon notes whether there is any obstacle to easy extension of the knees. If there should be (it is not often the case), an open incision must be made on each side of the popliteal space, and the tense hamstrings are in turn divided. If these incisions are long enough, the fascial contraction can be attacked on either side, for it is here that opposition is often found. I would discourage the use of a transverse incision, as when adopted it often seriously hampers the surgeon's efforts to fully extend the knee by reason of the strain cast upon the sutures. In 1885, when I was at the Stanley Hospital, there used to be an adult diplegic in a perambulator always at the gates, and on two or three occasions I took him in to try and straighten his contracted limbs. On one occasion I removed about an inch from each of the hamstrings, but he was mentally so deranged that we did not do each other any credit. I mention the fact, however, because Lorenz of Vienna has quite recently written on the advantage of exsecting portions of the hamstrings.

We have now presumably got our patient comfortably stretched upon an abduction frame, and we must keep him there for at least three months. Usually the wounds heal rapidly. Suppuration has occurred in the adductor cavity on three occasions only, despite the insanitary position of the wounds and the difficulty in adequately controlling the secretions. In 1890 I operated on twenty-seven patients, and this may be taken as a fair index of my yearly return.

At the end of three months the splint is taken off during the day and movements are sedulously practised. For some weeks stiffness exists, and often the movements are at first painful, but after a time, always shortened by vigorous exercise, the pain disappears and the effort must be made to walk.

The splints are of a simple kind, designed to keep the knee from bending. The boots should be made of felt with substantial soles. The nurse should be instructed to keep both boots and splints upon the patient day and night, and for the first two weeks, frequently during the day, abduction, adduction, flexion, and extension of the hip should be practised. This should be done with and without resistance. At night-time the feet should be attached to the side of the bed in order to maintain abduction. After the first few days of this later stage of treatment the splints should be removed twice a day and the muscles well massaged, and both active and passive movements of ankles, toes, knees and hips encouraged. Any movement executed in a jerky style should be practised until perfected.

The little patient may now try to walk. It will be noted that one of the difficulties of an untreated spastic when he tries to walk is the narrowing of the pedestal upon which the trunk rests by reason of the adducted limbs. Operation has now overcome this, and with abducted limbs the body is poised upon a widened pedestal. During early training the nurse must see that while walking the limbs do not approximate, and that, from the first, swinging of the limbs must be prohibited. Crutches should not be allowed until the patient has been taught to stand unsupported. I need not enter into any more detail regarding this most important stage of treatment, but would add that it affords an inexhaustible field for ingenuity, and that upon the intelligence and industry of the nurse very much depends.

Diplegic hands are treated on the same principles as I have enunciated in regard to infantile hemiplegia, and they must be trained to hold sticks and crutches with a firm unyielding grip.

I cannot now deal with individual cases, but I may say I have operated on cases from 12 months to 20 years of age. A large number of these were so bad that they had never attempted to place one foot before the other. Some were structurally flexed (contractured) at ankle, knee and hip. A most helpless youth of 20, one limb across the other, was able in six months to stand erect and walk with two sticks, and twelve months later could move his limbs north, south, east and west, with hardly an appreciable jerk. Success in an ancient case, where so much has to be unlearned, and where the mechanical stage offers such difficulty, proves the accuracy and efficacy of the principles I have endeavoured to expound. It is logical to infer that if old neglected cases are amenable to surgical education, that our prognosis should not be dismal in the young.

With regard to the degree of benefit to be derived from treatment, the parents should be given to understand that under favourable conditions of nursing and tuition the child, aided by the hand or sticks, will be able to walk varying distances in from twelve months to two years, and that with perfectly straight limbs and heels on *terra firma*. A large proportion of cases will later on manage aided by one stick. Even in the least successful cases, parents, mostly having despaired, are full of gratitude. The mental condition of the children obviously improves when their physical defects are remedied and they are enabled to mix with their little friends. Complete recovery in spastic paraplegia is of course impossible.

It will be gathered from my remarks that I wish to urge that the treatment of spastic paralysis should resolve itself into a system. Such a system involves operative, mechanical and educational stages. The treatment cannot be separated into parts. If the surgeon is not satisfied that the case is to be under his control for twelve months, he will consult his reputation best by leaving it alone. Operations not followed up by careful and prolonged after-care give rise to disappointment and discredit. Merely dividing tendons, to be followed by massage and electricity, is futile and dispiriting.

At the new Liverpool Country Hospital for Chronic Diseases of Children at Heswall, which we intend to build for 200 patients, we hope to have a ward for these paralytic cases, where we can keep them as long as needed. The nurses will be specially trained, and no opportunities will be lost from ignorance or neglect. The three months time limit which general hospitals impose materially blights the prospects of paralytic subjects. They are neglected at home, and wander from one institution to another, often the victims of conflicting theory and diverse practice. The successful hospital management of infantile paralysis is not complete without an organised system of education be inaugurated. This in the case of spastics must often be of the visual as opposed to the abstract type. Apart from its direct influence in improving the mind, we find it to have a sedative influence on the irritable. I have often thought that if there were scholastic institutes for the care of the paralytic children of the well-to-do, an undesirable gap would be filled. With cheerful surroundings, these little ones would enjoy in happy combination a development of both mind and body, under the scientific auspices of specially trained instructors. Such advantages would render material aid to many helpless little cripples. As it is, treatment is often half-hearted, rarely continuous, entered into with misgivings on the part of the surgeon and despair on the part of the patient's friends.

